CASE REPORT

# Didelphys uterus: misleading as ovarian cyst

S Nahar[1], B Das[2], F Begum[3]


## Abstract

*Mullerian duct anomalies are not uncommon. Anomalies may be diagnosed in infancy, adolescene or young adulthood. Female patient may present with a mass resulting from mucocolpos, haematocolpos, haematometra or primary amenorrhoea, delayed menarche, infertility and repeated pregnancy loses. A 14 years unmarried girl came with lower abdominal pain and mass with severe dysmenorrhoea with a history of lower abdominal surgery 5 months back. Clinically it was diagnosed as ovarian cyst but after laparotomy it was detected didelphys uterus. Left uterus was non-communicated with vagina forming haematometra. Right uterus well developed with one tube and healthy ovary.*


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## Case Report:

Miss Rima, age 14 yrs, a student of lower middle class family came from Chitolmari, Bagerhat, admitted to Khulna medical college on 27/09/10 with the complaints of cyclical pain in the left lower abdomen for 7 months; the pain was severe in nature. She had a history of abdominal surgery by MBBS doctor for ovarian cyst 5 months back without any document. On examination she is normotensive, mild anaemic & no other abnormality was detected.

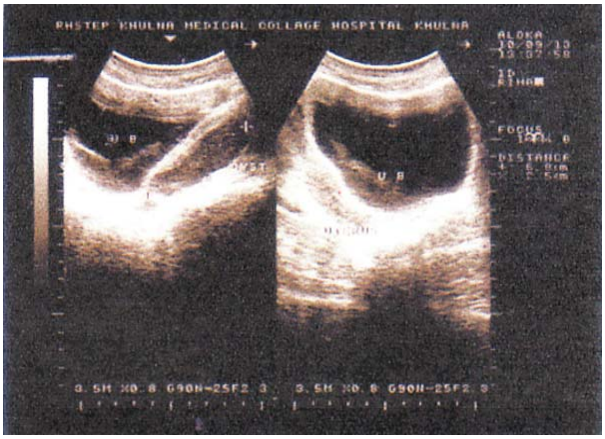


Fig 1 : Adenexae : A hypoechoic area about (68x25) mm is seen in adenexae with thick outline and no septation.

Per abdominal examination : A scar mark of lower transverse incision was seen & on palpation 6 X 5 cm mass was felt in the left illiac region which was firm in consistency & slightly moveable & tender and could not be well separated from the uterus. Per vaginal examination was same as per abdominal examination, through left fornix and other fornices were free & non tender.

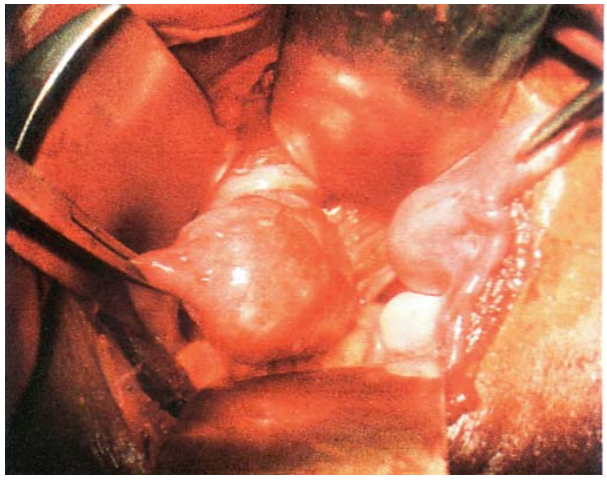

Fig 2 : Two uteri, connected with a transverse septum, right one well developed & left one enlarged.

Ultrasonography was done and the finding was a hypoechoic area about (68 X 25) mm is seen in left adenexa, no septation is seen within it, outline of the cystic mass is thick, No fluid is seen in the cul-de-sac. All investigations including Ca-125 were normal. After 3 weeks, laparotomy was done. There were two uteri with two cervices. The Right uterus was normal in size with one Fallopian tube and ovary. The left uterus was enlarged and there were no fallopian tube and ovary, probably due to prior operation. The cervix of the left uterus was closed, a firm cord like structure with non communication with the vagina. The left uterus was resected from its attachment. After operation, uterus was resected & old altered blood came out. On the 5th post operative day stitches were removed and the patient was discharged on that day.

## Introduction:

A congenital uterine malformation is a deviation from the shape or structure of a woman's own prenatal development. Incidence of congenital malformation reported in up to 3.2% of all women.1


1. Shamsun Nahar MS, Assoc. Professor, Department of Obs & Gynae, Khulna Medical College, Khulna.
2. Bishwajit Das DGO, Consultant (Gynae), Department of Obs & Gynae, Khulna Medical College, Khulna.
3. Ferdowsi Begum MBBS, Medical Officer (Gynae), Department of Obs & Gynae, Khulna Medical College, Khulna.

The actual incidence & prevalence of mullerian anomalies in the general population are unknown. Colombo reported the first documented case of vaginal agenesis in the 16th century.2 Incidence rate varies widely and depend on the study. Most author reported incidence of 0.1-3.5% in 2001.3,4 In women with infertility problem and recurrent abortion, the incidence of anomalies is slightly higher approximately 5-10%.5,6 Etiology of congenital malformation is sometimes categorized on the basis of genetic, environmental (So called multifactorial inheritance) factors. Two paired mullerian duct ultimately develop into the structure of the female reproductive tract. The ovaries & lower one third of the vagina has separate embryologic origin not derived from the Mullerian system.

Complete formation & differentiation of the Mullerian ducts into the segment of the female reproductive tract depend on completion of three phases of development as follows:

• Organogenesis: One or both Mullerian ducts may not develop fully, resulting in uterine agenesis or hypoplasia (bilateral) or unicornuate uterus (unilateral).

• Fusion: The process during which the lower segment of the paired Mullerian ducts fuse to form the uterus, cervix & upper vagina is termed lateral fusion. Failure in fusion result in anomalies such as bicornuate or didelphys uterus.

• Septal resorption: After the lower Mullerian ducts fuse, a central septum is present, which subsequently must be resorbed to form a single uterine cavity and cervix. Failure to resorption is the cause of septate uterus.

Mullerian duct anomalies are often associated with primary amenorrhea, treatable or untreatable causes of infertility, dysmenorrhea, dyspareunia, repeated first trimester spontaneous abortion, fetal intrauterine growth retardation, fetal malposition, preterm labour and retained placenta.7-9

## Discussion:

Didelphys uterus arises when midline fusion of the Mullerian ducts are arrested, either completely or incompletely. Approximately 11% of the uterine malformation is didelphys uterus.10 The complete form is characterized by two hemiuteri, two endocervical canals with cervices fused at the lower uterine segment. Each hemiuterus is associated with one fallopian tube. Ovarian malformation may also be present.11 The vagina may be single or double. Uterus didelphys is frequently associated with ipsilateral renal and ureter agenesis than any other type of Mullerian agenesis12. Our patient has two uterus, one Mullerian ducts failed to form complete uterus because whole body of uterus is well developed but cervix was thick cord like, no communication with vagina leads dysmenorrhea and haematometra after menarche.

Certain types of Mullerian duct anomaly can increase morbidity, such as patient with obstructed or partially obstructed Mullerian systems that present with haematosalpinx, haematocolpus, retrograde menses, and endometriosis. Anomalies may be diagnosed in infants, adolescences, or young adulthood. Female patients may present with mass resulting from an obstructed Mullerian system as infant (Mucocolpus), primary amenorrhea and mass (Haematocolpos) or delayed onset of menarche as adolescents or with problem of fertility and or carrying pregnancy to term as adults.13 Our patient presented with normal menstruation with severe dysmenorrhea. USG showing a cystic mass. This patient has a didelphys uterus but right horn is obstructed with functioning endometrium leads to haematometra. It may come to surgical attention when presented as an enlarging pelvic mass. If the contralateral healthy horn is almost fully developed, a full term pregnancy believes to be possible.14

Diagnosis of non obstructed uterine didelphys is usually asymptomatic until menarche. Only it can be detected during screening for causes of infertility or pregnancy loss. The diagnosis is often rendered during the initial pelvic examination when two cervices are identified. In obstructive type, the clinical presentations are variable and depend on the degree of obstruction and whether the obstruction has an opening or not. The most common presenting symptoms are onset of dysmenorrhea within the first years following menarche and progressive pelvic pain. A unilateral pelvic mass is detected on examination and presenting symptoms of marked rectal pain and constipation.

Diagnostic modalities are USG or TVS may be valuable adjunct. Other investigations include HSG, MRI and IVU to confirm and exclude associated urinary tract anomalies. MR1 is considered the criterion standard for imaging uterine anomalies. MRI provides high resolution images Of the uterine body, fundus & internal structures and it can help to evaluate the urinary tract for concomitant anomalies. In the past IVU was used for this purpose. Most types of uterine anomalies can be diagnosed by using MRI.15 MRI reveals two widely separated uterine horns and two cervices are typically identified. The intercornual angle is >60°. The zonal anatomy is presented within each hemiuterus. Surgical techniques for bicornuate uterus are metroplasty, the Strassmann procedure is the surgical treatment of choice for unifying the bicornuate and didelphys uteri. Transcervical lysis or transvaginal septum resections are used in other anomalies. In this patient we resected left sided haematometric mass. Her left tube

and ovary had resected by MBBS doctor previously due to missing diagnosis.Fig. l & 2 presented her USG and laparotomy finding.

Regarding her fertility & pregnancy outcome her right sided horn is completely developed with healthy tube and ovary. We counseled the patient about chance of preterm deliveries, live birth, ectopic pregnancies and spontaneous abortions. The poor reproductive outcomes are thought to be due to diminished uterine volumes and decreased perfusion of each hemiuterus.

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